

CHAPTER XVII

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Congenital malformations, deformations and chromosomal abnormalities (Q00-Q99)

Excludes1: inborn errors of metabolism (E70-E90)

This chapter contains the following blocks:

- Q00-Q07 Congenital malformations of the nervous system
- Q10-Q18 Congenital malformations of eye, ear, face and neck
- Q20-Q28 Congenital malformations of the circulatory system
- Q30-Q34 Congenital malformations of the respiratory system
- Q35-Q37 Cleft lip and cleft palate
- Q38-Q45 Other congenital malformations of the digestive system
- Q50-Q56 Congenital malformations of genital organs
- Q60-Q64 Congenital malformations of the urinary system
- Q65-Q79 Congenital malformations and deformations of the musculoskeletal system
- Q80-Q89 Other congenital malformations
- Q90-Q99 Chromosomal abnormalities, not elsewhere classified

Congenital malformations of the nervous system (Q00-Q07)

Q00 Anencephaly and similar malformations

Q00.0 Anencephaly

- Acephaly
- Acrania
- Amyelencephaly
- Hemianencephaly
- Hemicephaly

Q00.1 Craniorachischisis

Q00.2 Iniencephaly

Q01 Encephalocele

- Includes:
- Arnold-Chiari syndrome, type III
 - encephalocystocele
 - encephalomylomeningocele
 - hydroencephalocele
 - hydromeningocele, cranial
 - meningocele, cerebral

meningoencephalocele
Excludes1: Meckel-Gruber syndrome (Q61.9)

- Q01.0 Frontal encephalocele**
- Q01.1 Nasofrontal encephalocele**
- Q01.2 Occipital encephalocele**
- Q01.8 Encephalocele of other sites**
- Q01.9 Encephalocele, unspecified**

Q02 Microcephaly

Includes: hydromicrocephaly
micrencephalon
Excludes1: Meckel-Gruber syndrome (Q61.9)

Q03 Congenital hydrocephalus

Includes: hydrocephalus in newborn
Excludes1: Arnold-Chiari syndrome, type II (Q07.0-07.9)
acquired hydrocephalus (G91.-)
hydrocephalus due to congenital toxoplasmosis (P37.1)
hydrocephalus with spina bifida (Q05.0-Q05.4)

Q03.0 Malformations of aqueduct of Sylvius

Anomaly of aqueduct of Sylvius
Obstruction of aqueduct of Sylvius, congenital
Stenosis of aqueduct of Sylvius

- Q03.1 Atresia of foramina of Magendie and Luschka**
Dandy-Walker syndrome
- Q03.8 Other congenital hydrocephalus**
- Q03.9 Congenital hydrocephalus, unspecified**

Q04 Other congenital malformations of brain

Excludes1: cyclopia (Q87.0)
macrocephaly (Q75.3)

Q04.0 Congenital malformations of corpus callosum

Agenesis of corpus callosum

Q04.1 Arhinencephaly

Q04.2 Holoprosencephaly

Q04.3 Other reduction deformities of brain

Absence of part of brain

Agenesis of part of brain

Agyria

Aplasia of part of brain

Hydranencephaly

Hypoplasia of part of brain

- Lissencephaly
Microgyria
Pachygyria
Excludes1: congenital malformations of corpus callosum (Q04.0)
- Q04.4 Septo-optic dysplasia of brain**
- Q04.5 Megalencephaly**
- Q04.6 Congenital cerebral cysts**
- Porencephaly
Schizencephaly
Excludes1: acquired porencephalic cyst (G93.0)
- Q04.8 Other specified congenital malformations of brain**
- Arnold-Chiari syndrome, type IV
Macrogyria
- Q04.9 Congenital malformation of brain, unspecified**
- Congenital anomaly NOS of brain
Congenital deformity NOS of brain
Congenital disease or lesion NOS of brain
Multiple anomalies NOS of brain, congenital

Q05 Spina bifida

- Includes: hydromeningocele (spinal)
meningocele (spinal)
meningomyelocele
myelocele
myelomeningocele
rachischisis
spina bifida (aperta)(cystica)
syringomyelocele
- Excludes1: Arnold-Chiari syndrome, type II (Q07.0-)
spina bifida occulta (Q76.0)

- Q05.0 Cervical spina bifida with hydrocephalus**
- Q05.1 Thoracic spina bifida with hydrocephalus**
- Dorsal spina bifida with hydrocephalus
Thoracolumbar spina bifida with hydrocephalus
- Q05.2 Lumbar spina bifida with hydrocephalus**
- Lumbosacral spina bifida with hydrocephalus
- Q05.3 Sacral spina bifida with hydrocephalus**
- Q05.4 Unspecified spina bifida with hydrocephalus**
- Q05.5 Cervical spina bifida without hydrocephalus**
- Q05.6 Thoracic spina bifida without hydrocephalus**
- Dorsal spina bifida NOS
Thoracolumbar spina bifida NOS
- Q05.7 Lumbar spina bifida without hydrocephalus**
- Lumbosacral spina bifida NOS
- Q05.8 Sacral spina bifida without hydrocephalus**

Q05.9 Spina bifida, unspecified

Q06 Other congenital malformations of spinal cord

Q06.0 Amyelia

Q06.1 Hypoplasia and dysplasia of spinal cord

Atelomyelia

Myelatelia

Myelodysplasia of spinal cord

Q06.2 Diastematomyelia

Q06.3 Other congenital cauda equina malformations

Q06.4 Hydromyelia

Hydrorachis

Q06.8 Other specified congenital malformations of spinal cord

Q06.9 Congenital malformation of spinal cord, unspecified

Congenital anomaly NOS of spinal cord

Congenital deformity NOS of spinal cord

Congenital disease or lesion NOS of spinal cord

Q07 Other congenital malformations of nervous system

Excludes1: familial dysautonomia [Riley-Day] (G90.1)

neurofibromatosis (nonmalignant) (Q85.0)

Q07.0 Arnold-Chiari syndrome

Arnold-Chiari syndrome, type II

Excludes1: Arnold-Chiari syndrome, type III (Q01.-)

Arnold-Chiari syndrome, type IV (Q04.8)

Q07.00 Arnold-Chiari syndrome without spina bifida or hydrocephalus

Q07.01 Arnold-Chiari syndrome with spina bifida

Q07.02 Arnold-Chiari syndrome with hydrocephalus

Q07.03 Arnold-Chiari syndrome with spina bifida and hydrocephalus

Q07.8 Other specified congenital malformations of nervous system

Agenesis of nerve

Displacement of brachial plexus

Jaw-winking syndrome

Marcus Gunn's syndrome

Q07.9 Congenital malformation of nervous system, unspecified

Congenital anomaly NOS of nervous system

Congenital deformity NOS of nervous system

Congenital disease or lesion NOS of nervous system

Congenital malformations of eye, ear, face and neck

(Q10-Q18)

Excludes2: cleft lip and cleft palate (Q35-Q37)

congenital malformation of:

cervical spine (Q05.0, Q05.5, Q67.5, Q76.0-Q76.4)
larynx (Q31.-)
lip NEC (Q38.0)
nose (Q30.-)
parathyroid gland (Q89.2)
thyroid gland (Q89.2)

Q10 Congenital malformations of eyelid, lacrimal apparatus and orbit

Excludes1: cryptophthalmos NOS (Q11.2)
cryptophthalmos syndrome (Q87.0)

Q10.0 Congenital ptosis

Q10.1 Congenital ectropion

Q10.2 Congenital entropion

Q10.3 Other congenital malformations of eyelid

Ablepharon

Blepharophimosis, congenital

Coloboma of eyelid

Congenital absence or agenesis of cilia

Congenital absence or agenesis of eyelid

Congenital accessory eyelid

Congenital accessory eye muscle

Congenital malformation of eyelid NOS

Q10.4 Absence and agenesis of lacrimal apparatus

Congenital absence of punctum lacrimale

Q10.5 Congenital stenosis and stricture of lacrimal duct

Q10.6 Other congenital malformations of lacrimal apparatus

Congenital malformation of lacrimal apparatus NOS

Q10.7 Congenital malformation of orbit

Q11 Anophthalmos, microphthalmos and macrophtalmos

Q11.0 Cystic eyeball

Q11.1 Other anophthalmos

Anophthalmos NOS

Agenesis of eye

Aplasia of eye

Q11.2 Microphthalmos

Cryptophthalmos NOS

Dysplasia of eye

Hypoplasia of eye

Rudimentary eye

Excludes1: cryptophthalmos syndrome (Q87.0)

Q11.3 Macrophtalmos

Excludes1: macrophtalmos in congenital glaucoma (Q15.0)

Q12 Congenital lens malformations

- Q12.0 Congenital cataract**
- Q12.1 Congenital displaced lens**
- Q12.2 Coloboma of lens**
- Q12.3 Congenital aphakia**
- Q12.4 Spherophakia**
- Q12.8 Other congenital lens malformations**
 - Microphtalmia
- Q12.9 Congenital lens malformation, unspecified**

Q13 Congenital malformations of anterior segment of eye

- Q13.0 Coloboma of iris**
 - Coloboma NOS
- Q13.1 Absence of iris**
 - Aniridia
 - Use additional code for associated glaucoma (H42)
- Q13.2 Other congenital malformations of iris**
 - Anisocoria, congenital
 - Atresia of pupil
 - Congenital malformation of iris NOS
 - Corectopia
- Q13.3 Congenital corneal opacity**
- Q13.4 Other congenital corneal malformations**
 - Congenital malformation of cornea NOS
 - Microcornea
 - Peter's anomaly
- Q13.5 Blue sclera**
- Q13.8 Other congenital malformations of anterior segment of eye**
 - Q13.81 Rieger's anomaly**
 - Use additional code for associated glaucoma (H42)
 - Q13.89 Other congenital malformations of anterior segment of eye**
- Q13.9 Congenital malformation of anterior segment of eye, unspecified**

Q14 Congenital malformations of posterior segment of eye

- Q14.0 Congenital malformation of vitreous humor**
 - Congenital vitreous opacity
- Q14.1 Congenital malformation of retina**
 - Congenital retinal aneurysm
- Q14.2 Congenital malformation of optic disc**
 - Coloboma of optic disc
- Q14.3 Congenital malformation of choroid**
- Q14.8 Other congenital malformations of posterior segment of eye**

Coloboma of the fundus
Q14.9 Congenital malformation of posterior segment of eye, unspecified

Q15 Other congenital malformations of eye

Excludes1: congenital nystagmus (H55.01)
ocular albinism (E70.31-)
retinitis pigmentosa (H35.52)

Q15.0 Congenital glaucoma

Axenfeld's anomaly
Buphthalmos
Glaucoma of childhood
Glaucoma of newborn
Hydrophthalmos
Keratoglobus, congenital
Macrophtalmos in congenital glaucoma
Megalocornea

Q15.8 Other specified congenital malformations of eye

Q15.9 Congenital malformation of eye, unspecified

Congenital anomaly of eye
Congenital deformity of eye

Q16 Congenital malformations of ear causing impairment of hearing

Excludes1: congenital deafness (H90.-)

Q16.0 Congenital absence of (ear) auricle

Q16.1 Congenital absence, atresia and stricture of auditory canal (external)
Congenital atresia or stricture of osseous meatus

Q16.2 Absence of eustachian tube

Q16.3 Congenital malformation of ear ossicles
Congenital fusion of ear ossicles

Q16.4 Other congenital malformations of middle ear

Congenital malformation of middle ear NOS

Q16.5 Congenital malformation of inner ear

Congenital anomaly of membranous labyrinth
Congenital anomaly of organ of Corti

Q16.9 Congenital malformation of ear causing impairment of hearing, unspecified
Congenital absence of ear NOS

Q17 Other congenital malformations of ear

Excludes1: congenital malformations of ear with impairment of hearing (Q16.0- Q16.9)
preauricular sinus (Q18.1)

Q17.0 Accessory auricle

Accessory tragus

- Polyotia
- Preauricular appendage or tag
- Supernumerary ear
- Supernumerary lobule
- Q17.1 Macrotia**
- Q17.2 Microtia**
- Q17.3 Other misshapen ear**
 - Pointed ear
- Q17.4 Misplaced ear**
 - Low-set ears
 - Excludes1: cervical auricle (Q18.2)
- Q17.5 Prominent ear**
 - Bat ear
- Q17.8 Other specified congenital malformations of ear**
 - Congenital absence of lobe of ear
- Q17.9 Congenital malformation of ear, unspecified**
 - Congenital anomaly of ear NOS

Q18 Other congenital malformations of face and neck

Excludes1: cleft lip and cleft palate (Q35-Q37)
conditions classified to Q67.0-Q67.4
congenital malformations of skull and face bones (Q75.-)
cyclopia (Q87.0)
dentofacial anomalies [including malocclusion] (M26.-)
malformation syndromes affecting facial appearance (Q87.0)
persistent thyroglossal duct (Q89.2)

- Q18.0 Sinus, fistula and cyst of branchial cleft**
 - Branchial vestige
- Q18.1 Preauricular sinus and cyst**
 - Fistula of auricle, congenital
 - Cervicoaural fistula
- Q18.2 Other branchial cleft malformations**
 - Branchial cleft malformation NOS
 - Cervical auricle
 - Otocephaly
- Q18.3 Webbing of neck**
 - Pterygium colli
- Q18.4 Macrostomia**
- Q18.5 Microstomia**
- Q18.6 Macrocheilia**
 - Hypertrophy of lip, congenital
- Q18.7 Microcheilia**
- Q18.8 Other specified congenital malformations of face and neck**
 - Medial cyst of face and neck
 - Medial fistula of face and neck

- Medial sinus of face and neck
- Q18.9 Congenital malformation of face and neck, unspecified**
- Congenital anomaly NOS of face and neck

Congenital malformations of the circulatory system (Q20-Q28)

Q20 Congenital malformations of cardiac chambers and connections

Excludes1: dextrocardia with situs inversus (Q89.3)
mirror-image atrial arrangement with situs inversus (Q89.3)

Q20.0 Common arterial trunk

Persistent truncus arteriosus

Excludes1: aortic septal defect (Q21.4)

Q20.1 Double outlet right ventricle

Taussig-Bing syndrome

Q20.2 Double outlet left ventricle

Q20.3 Discordant ventriculoarterial connection

Dextrotransposition of aorta

Transposition of great vessels (complete)

Q20.4 Double inlet ventricle

Common ventricle

Cor triloculare biventriculare

Single ventricle

Q20.5 Discordant atrioventricular connection

Corrected transposition

Levotransposition

Ventricular inversion

Q20.6 Isomerism of atrial appendages

Isomerism of atrial appendages with asplenia or polysplenia

Q20.8 Other congenital malformations of cardiac chambers and connections

Cor binoculare

Q20.9 Congenital malformation of cardiac chambers and connections, unspecified

Q21 Congenital malformations of cardiac septa

Excludes1: acquired cardiac septal defect (I51.0)

Q21.0 Ventricular septal defect

Roger's disease

Q21.1 Atrial septal defect

Coronary sinus defect

Patent or persistent foramen ovale

Patent or persistent ostium secundum defect (type II)

Patent or persistent sinus venosus defect

- Q21.2 Atrioventricular septal defect**
Common atrioventricular canal
Endocardial cushion defect
Ostium primum atrial septal defect (type I)
- Q21.3 Tetralogy of Fallot**
Ventricular septal defect with pulmonary stenosis or atresia, dextroposition of aorta and hypertrophy of right ventricle.
- Q21.4 Aortopulmonary septal defect**
Aortic septal defect
Aortopulmonary window
- Q21.8 Other congenital malformations of cardiac septa**
Eisenmenger's syndrome
Pentalogy of Fallot
- Q21.9 Congenital malformation of cardiac septum, unspecified**
Septal (heart) defect NOS

Q22 Congenital malformations of pulmonary and tricuspid valves

- Q22.0 Pulmonary valve atresia**
- Q22.1 Congenital pulmonary valve stenosis**
- Q22.2 Congenital pulmonary valve insufficiency**
Congenital pulmonary valve regurgitation
- Q22.3 Other congenital malformations of pulmonary valve**
Congenital malformation of pulmonary valve NOS
Supernumerary cusps of pulmonary valve
- Q22.4 Congenital tricuspid stenosis**
Congenital tricuspid atresia
- Q22.5 Ebstein's anomaly**
- Q22.6 Hypoplastic right heart syndrome**
- Q22.8 Other congenital malformations of tricuspid valve**
- Q22.9 Congenital malformation of tricuspid valve, unspecified**

Q23 Congenital malformations of aortic and mitral valves

- Q23.0 Congenital stenosis of aortic valve**
Congenital aortic atresia
Congenital aortic stenosis NOS
Excludes1: congenital stenosis of aortic valve in hypoplastic left heart syndrome (Q23.4)
congenital subaortic stenosis (Q24.4)
supravalvular aortic stenosis (congenital) (Q25.3)
- Q23.1 Congenital insufficiency of aortic valve**
Bicuspid aortic valve
Congenital aortic insufficiency
- Q23.2 Congenital mitral stenosis**
Congenital mitral atresia

- Q23.3** Congenital mitral insufficiency
- Q23.4** Hypoplastic left heart syndrome
- Q23.8** Other congenital malformations of aortic and mitral valves
- Q23.9** Congenital malformation of aortic and mitral valves, unspecified

Q24 Other congenital malformations of heart

Excludes1: endocardial fibroelastosis (I42.4)

Q24.0 Dextrocardia

Excludes1: dextrocardia with situs inversus (Q89.3)
isomerism of atrial appendages (with asplenia or polysplenia) (Q20.6)
mirror-image atrial arrangement with situs inversus (Q89.3)

Q24.1 Levocardia

Q24.2 Cor triatriatum

Q24.3 Pulmonary infundibular stenosis

Subvalvular pulmonic stenosis

Q24.4 Congenital subaortic stenosis

Q24.5 Malformation of coronary vessels

Congenital coronary (artery) aneurysm

Q24.6 Congenital heart block

Q24.8 Other specified congenital malformations of heart

Congenital diverticulum of left ventricle

Congenital malformation of myocardium

Congenital malformation of pericardium

Malposition of heart

Uhl's disease

Q24.9 Congenital malformation of heart, unspecified

Congenital anomaly of heart

Congenital disease of heart

Q25 Congenital malformations of great arteries

Q25.0 Patent ductus arteriosus

Patent ductus Botallo

Persistent ductus arteriosus

Q25.1 Coarctation of aorta

Coarctation of aorta (preductal) (postductal)

Q25.2 Atresia of aorta

Q25.3 Supravalvular aortic stenosis

Excludes1: congenital aortic stenosis NOS (Q23.0)

congenital aortic valve stenosis (Q23.0)

Q25.4 Other congenital malformations of aorta

Absence of aorta

Aneurysm of sinus of Valsalva (ruptured)

Aplasia of aorta

- Congenital aneurysm of aorta
- Congenital malformations of aorta
- Congenital dilatation of aorta
- Double aortic arch [vascular ring of aorta]
- Hypoplasia of aorta
- Persistent convolutions of aortic arch
- Persistent right aortic arch
- Excludes1: hypoplasia of aorta in hypoplastic left heart syndrome (Q23.4)
- Q25.5 Atresia of pulmonary artery**
- Q25.6 Stenosis of pulmonary artery**
- Q25.7 Other congenital malformations of pulmonary artery**
 - Aberrant pulmonary artery
 - Agenesis of pulmonary artery
 - Congenital aneurysm of pulmonary artery
 - Congenital anomaly of pulmonary artery
 - Congenital pulmonary arteriovenous aneurysm
 - Hypoplasia of pulmonary artery
- Q25.8 Other congenital malformations of other great arteries**
- Q25.9 Congenital malformation of great arteries, unspecified**

Q26 Congenital malformations of great veins

- Q26.0 Congenital stenosis of vena cava**
 - Congenital stenosis of vena cava (inferior)(superior)
- Q26.1 Persistent left superior vena cava**
- Q26.2 Total anomalous pulmonary venous connection**
 - Total anomalous pulmonary venous return [TAPVR], subdiaphragmatic
 - Total anomalous pulmonary venous return [TAPVR], supradiaphragmatic
- Q26.3 Partial anomalous pulmonary venous connection**
 - Partial anomalous pulmonary venous return
- Q26.4 Anomalous pulmonary venous connection, unspecified**
- Q26.5 Anomalous portal venous connection**
- Q26.6 Portal vein-hepatic artery fistula**
- Q26.8 Other congenital malformations of great veins**
 - Absence of vena cava (inferior) (superior)
 - Azygos continuation of inferior vena cava
 - Persistent left posterior cardinal vein
 - Scimitar syndrome
- Q26.9 Congenital malformation of great vein, unspecified**
 - Congenital anomaly of vena cava (inferior) (superior) NOS

Q27 Other congenital malformations of peripheral vascular system

- Excludes2: anomalies of cerebral and precerebral vessels (Q28.0-Q28.3)
 - anomalies of coronary vessels (Q24.5)
 - anomalies of pulmonary artery (Q25.5-Q25.7)
 - congenital retinal aneurysm (Q14.1)

hemangioma and lymphangioma (D18.-)

- Q27.0 Congenital absence and hypoplasia of umbilical artery**
Single umbilical artery
- Q27.1 Congenital renal artery stenosis**
- Q27.2 Other congenital malformations of renal artery**
Congenital malformation of renal artery NOS
Multiple renal arteries
- Q27.3 Arteriovenous malformation (peripheral)**
Arteriovenous aneurysm
Excludes1: acquired arteriovenous aneurysm (I77.0)
Excludes2: arteriovenous malformation of cerebral vessels (Q28.2)
arteriovenous malformation of precerebral vessels (Q28.0)
- Q27.30 Arteriovenous malformation, site unspecified**
- Q27.31 Arteriovenous malformation of vessel of upper limb**
- Q27.32 Arteriovenous malformation of vessel of lower limb**
- Q27.33 Arteriovenous malformation of digestive system vessel**
- Q27.34 Arteriovenous malformation of renal vessel**
- Q27.39 Arteriovenous malformation, other site**
- Q27.4 Congenital phlebectasia**
- Q27.8 Other specified congenital malformations of peripheral vascular system**
Absence of peripheral vascular system
Atresia of peripheral vascular system
Congenital aneurysm (peripheral)
Congenital stricture, artery
Congenital varix
Excludes1: arteriovenous malformation (Q27.3-)
- Q27.9 Congenital malformation of peripheral vascular system, unspecified**
Anomaly of artery or vein NOS

Q28 Other congenital malformations of circulatory system

- Excludes1: congenital aneurysm NOS (Q27.8)
congenital coronary aneurysm (Q24.5)
ruptured cerebral arteriovenous malformation (I60.8)
ruptured malformation of precerebral vessels (I72.0)
- Excludes2: congenital peripheral aneurysm (Q27.8)
congenital pulmonary aneurysm (Q25.7)
congenital retinal aneurysm (Q14.1)

- Q28.0 Arteriovenous malformation of precerebral vessels**
Congenital arteriovenous precerebral aneurysm (nonruptured)
- Q28.1 Other malformations of precerebral vessels**
Congenital malformation of precerebral vessels NOS
Congenital precerebral aneurysm (nonruptured)
- Q28.2 Arteriovenous malformation of cerebral vessels**
Arteriovenous malformation of brain NOS

- Congenital arteriovenous cerebral aneurysm (nonruptured)
- Q28.3 Other malformations of cerebral vessels**
 - Congenital cerebral aneurysm (nonruptured)
 - Congenital malformation of cerebral vessels NOS
- Q28.8 Other specified congenital malformations of circulatory system**
 - Congenital aneurysm, specified site NEC
 - Spinal vessel anomaly
- Q28.9 Congenital malformation of circulatory system, unspecified**

Congenital malformations of the respiratory system (Q30-Q34)

Q30 Congenital malformations of nose

Excludes1: congenital deviation of nasal septum (Q67.4)

- Q30.0 Choanal atresia**
 - Atresia of nares (anterior) (posterior)
 - Congenital stenosis of nares (anterior) (posterior)
- Q30.1 Agenesis and underdevelopment of nose**
 - Congenital absent of nose
- Q30.2 Fissured, notched and cleft nose**
- Q30.3 Congenital perforated nasal septum**
- Q30.8 Other congenital malformations of nose**
 - Accessory nose
 - Congenital anomaly of nasal sinus wall
- Q30.9 Congenital malformation of nose, unspecified**

Q31 Congenital malformations of larynx

- Q31.0 Web of larynx**
 - Glottic web of larynx
 - Subglottic web of larynx
 - Web of larynx NOS
- Q31.1 Congenital subglottic stenosis**
- Q31.2 Laryngeal hypoplasia**
- Q31.3 Laryngocoele**
- Q31.4 Congenital laryngeal stridor**
 - Congenital stridor (larynx) NOS
- Q31.8 Other congenital malformations of larynx**
 - Absence of larynx
 - Agenesis of larynx
 - Atresia of larynx
 - Congenital cleft thyroid cartilage
 - Congenital fissure of epiglottis

Congenital stenosis of larynx NEC
Posterior cleft of cricoid cartilage
Q31.9 Congenital malformation of larynx, unspecified

Q32 Congenital malformations of trachea and bronchus

Excludes1: congenital bronchiectasis (Q33.4)

Q32.0 Congenital tracheomalacia

Q32.1 Other congenital malformations of trachea

Atresia of trachea
Congenital anomaly of tracheal cartilage
Congenital dilatation of trachea
Congenital malformation of trachea
Congenital stenosis of trachea
Congenital tracheocele

Q32.2 Congenital bronchomalacia

Q32.3 Congenital stenosis of bronchus

Q32.8 Other congenital malformations of bronchus

Absence of bronchus
Agenesis of bronchus
Atresia of bronchus
Congenital diverticulum of bronchus
Congenital malformation of bronchus NOS

Q33 Congenital malformations of lung

Q33.0 Congenital cystic lung

Congenital cystic lung disease
Congenital honeycomb lung
Congenital polycystic lung disease
Excludes1: cystic fibrosis (E84.0)
cystic lung disease, acquired or unspecified (J98.4)

Q33.1 Accessory lobe of lung

Azygos lobe (fissured), lung

Q33.2 Sequestration of lung

Q33.3 Congenital agenesis of lung

Congenital absence of lung (lobe)

Q33.4 Congenital bronchiectasis

Q33.5 Ectopic tissue in lung

Q33.6 Congenital hypoplasia and dysplasia of lung

Excludes1: pulmonary hypoplasia associated with short gestation (P28.0)

Q33.8 Other congenital malformations of lung

Q33.9 Congenital malformation of lung, unspecified

Q34 Other congenital malformations of respiratory system

- Q34.0 Anomaly of pleura**
- Q34.1 Congenital cyst of mediastinum**
- Q34.8 Other specified congenital malformations of respiratory system**
 - Atresia of nasopharynx
- Q34.9 Congenital malformation of respiratory system, unspecified**
 - Congenital absence of respiratory system
 - Congenital anomaly of respiratory system NOS

Cleft lip and cleft palate

(Q35-Q37)

Excludes1: Robin's syndrome (Q87.0)

Q35 Cleft palate

Includes: fissure of palate
palatoschisis

Excludes1: cleft palate with cleft lip (Q37.-)

- Q35.1 Cleft hard palate**
- Q35.3 Cleft soft palate**
- Q35.5 Cleft hard palate with cleft soft palate**
- Q35.6 Cleft palate, medial**
- Q35.7 Cleft uvula**
- Q35.9 Cleft palate, unspecified**
 - Cleft palate NOS

Q36 Cleft lip

Includes: cheiloschisis
congenital fissure of lip
harelip
labium leporinum

Excludes1: cleft lip with cleft palate (Q37.-)

- Q36.0 Cleft lip, bilateral**
- Q36.1 Cleft lip, medial**
- Q36.9 Cleft lip, unilateral**
 - Cleft lip NOS

Q37 Cleft palate with cleft lip

Includes: cheilopalatoschisis

- Q37.0 Cleft hard palate with bilateral cleft lip**
- Q37.1 Cleft hard palate with unilateral cleft lip**
 - Cleft hard palate with cleft lip NOS

- Q37.2 Cleft soft palate with bilateral cleft lip**
- Q37.3 Cleft soft palate with unilateral cleft lip**
 - Cleft soft palate with cleft lip NOS
- Q37.4 Cleft hard and soft palate with bilateral cleft lip**
- Q37.5 Cleft hard and soft palate with unilateral cleft lip**
 - Cleft hard and soft palate with cleft lip NOS
- Q37.8 Unspecified cleft palate with bilateral cleft lip**
- Q37.9 Unspecified cleft palate with unilateral cleft lip**
 - Cleft palate with cleft lip NOS

Other congenital malformations of the digestive system (Q38-Q45)

Q38 Other congenital malformations of tongue, mouth and pharynx

Excludes1: dentofacial anomalies (M26.-)
macrostomia (Q18.4)
microstomia (Q18.5)

Q38.0 Congenital malformations of lips, not elsewhere classified

Congenital fistula of lip
Congenital malformation of lip NOS
Van der Woude's syndrome
Excludes1:
cleft lip (Q36.-)
cleft lip with cleft palate (Q37.-)
macrocheilia (Q18.6)
microcheilia (Q18.7)

Q38.1 Ankyloglossia

Tongue tie

Q38.2 Macroglossia

Congenital hypertrophy of tongue

Q38.3 Other congenital malformations of tongue

Aglossia
Bifid tongue
Congenital adhesion of tongue
Congenital fissure of tongue
Congenital malformation of tongue NOS
Double tongue
Hypoglossia
Hypoplasia of tongue
Microglossia

Q38.4 Congenital malformations of salivary glands and ducts

Atresia of salivary glands and ducts
Congenital absence of salivary glands and ducts
Congenital accessory salivary glands and ducts

- Congenital fistula of salivary gland
- Q38.5 Congenital malformations of palate, not elsewhere classified**
 - Congenital absence of uvula
 - Congenital malformation of palate NOS
 - Congenital high arched palate
 - Excludes1: cleft palate (Q35.-)
 - cleft palate with cleft lip (Q37.-)
- Q38.6 Other congenital malformations of mouth**
 - Congenital malformation of mouth NOS
- Q38.7 Congenital pharyngeal pouch**
 - Congenital diverticulum of pharynx
 - Excludes1: pharyngeal pouch syndrome (D82.1)
- Q38.8 Other congenital malformations of pharynx**
 - Congenital malformation of pharynx NOS
 - Imperforate pharynx

Q39 Congenital malformations of esophagus

- Q39.0 Atresia of esophagus without fistula**
 - Atresia of esophagus NOS
- Q39.1 Atresia of esophagus with tracheo-esophageal fistula**
 - Atresia of esophagus with broncho-esophageal fistula
- Q39.2 Congenital tracheo-esophageal fistula without atresia**
 - Congenital tracheo-esophageal fistula NOS
- Q39.3 Congenital stenosis and stricture of esophagus**
- Q39.4 Esophageal web**
- Q39.5 Congenital dilatation of esophagus**
- Q39.6 Congenital diverticulum of esophagus**
 - Congenital esophageal pouch
- Q39.8 Other congenital malformations of esophagus**
 - Congenital absence of esophagus
 - Congenital displacement of esophagus
 - Congenital duplication of esophagus
- Q39.9 Congenital malformation of esophagus, unspecified**

Q40 Other congenital malformations of upper alimentary tract

- Q40.0 Congenital hypertrophic pyloric stenosis**
 - Congenital or infantile constriction
 - Congenital or infantile hypertrophy
 - Congenital or infantile spasm
 - Congenital or infantile stenosis
 - Congenital or infantile stricture
- Q40.1 Congenital hiatus hernia**
 - Congenital displacement of cardia through esophageal hiatus
 - Excludes1: congenital diaphragmatic hernia (Q79.0)

- Q40.2 Other specified congenital malformations of stomach**
- Congenital cardiospasm
 - Congenital displacement of stomach
 - Congenital diverticulum of stomach
 - Congenital hourglass stomach
 - Congenital duplication of stomach
 - Megalogastria
 - Microgastria
- Q40.3 Congenital malformation of stomach, unspecified**
- Q40.8 Other specified congenital malformations of upper alimentary tract**
- Q40.9 Congenital malformation of upper alimentary tract, unspecified**
- Congenital anomaly of upper alimentary tract
 - Congenital deformity of upper alimentary tract

Q41 Congenital absence, atresia and stenosis of small intestine

Includes: congenital obstruction, occlusion or stricture of small intestine or intestine NOS

Excludes1: cystic fibrosis with intestinal manifestation (E84.1)
meconium ileus (P75)

- Q41.0 Congenital absence, atresia and stenosis of duodenum**
- Q41.1 Congenital absence, atresia and stenosis of jejunum**
- Apple peel syndrome
 - Imperforate jejunum
- Q41.2 Congenital absence, atresia and stenosis of ileum**
- Q41.8 Congenital absence, atresia and stenosis of other specified parts of small intestine**
- Q41.9 Congenital absence, atresia and stenosis of small intestine, part unspecified**
- Congenital absence, atresia and stenosis of intestine NOS

Q42 Congenital absence, atresia and stenosis of large intestine

Includes: congenital obstruction, occlusion and stricture of large intestine

- Q42.0 Congenital absence, atresia and stenosis of rectum with fistula**
- Q42.1 Congenital absence, atresia and stenosis of rectum without fistula**
- Imperforate rectum
- Q42.2 Congenital absence, atresia and stenosis of anus with fistula**
- Q42.3 Congenital absence, atresia and stenosis of anus without fistula**
- Imperforate anus
- Q42.8 Congenital absence, atresia and stenosis of other parts of large intestine**
- Q42.9 Congenital absence, atresia and stenosis of large intestine, part unspecified**

Q43 Other congenital malformations of intestine

- Q43.0 Meckel's diverticulum (displaced) (hypertrophic)**

- Persistent omphalomesenteric duct
Persistent vitelline duct
- Q43.1 Hirschsprung's disease**
Aganglionosis
Congenital (aganglionic) megacolon
- Q43.2 Other congenital functional disorders of colon**
Congenital dilatation of colon
- Q43.3 Congenital malformations of intestinal fixation**
Congenital omental, anomalous adhesions [bands]
Congenital peritoneal adhesions [bands]
Incomplete rotation of cecum and colon
Insufficient rotation of cecum and colon
Jackson's membrane
Malrotation of colon
Rotation failure of cecum and colon
Universal mesentery
- Q43.4 Duplication of intestine**
- Q43.5 Ectopic anus**
- Q43.6 Congenital fistula of rectum and anus**
Excludes1: congenital fistula of anus with absence, atresia and stenosis
 (Q42.2)
congenital fistula of rectum with absence, atresia and stenosis
 (Q42.0)
congenital rectovaginal fistula (Q52.2)
congenital urethrorectal fistula (Q64.7)
pilonidal fistula or sinus (L05.-)
- Q43.7 Persistent cloaca**
Cloaca NOS
- Q43.8 Other specified congenital malformations of intestine**
Congenital blind loop syndrome
Congenital diverticulitis, colon
Congenital diverticulum, intestine
Dolichocolon
Megalappendix
Megaloduodenum
Microcolon
Transposition of appendix
Transposition of colon
Transposition of intestine
- Q43.9 Congenital malformation of intestine, unspecified**

Q44 Congenital malformations of gallbladder, bile ducts and liver

- Q44.0 Agenesis, aplasia and hypoplasia of gallbladder**
Congenital absence of gallbladder
- Q44.1 Other congenital malformations of gallbladder**

- Congenital malformation of gallbladder NOS
- Intrahepatic gallbladder
- Q44.2 Atresia of bile ducts**
- Q44.3 Congenital stenosis and stricture of bile ducts**
- Q44.4 Choledochal cyst**
- Q44.5 Other congenital malformations of bile ducts**
 - Accessory hepatic duct
 - Biliary duct duplication
 - Congenital malformation of bile duct NOS
 - Cystic duct duplication
- Q44.6 Cystic disease of liver**
 - Fibrocystic disease of liver
- Q44.7 Other congenital malformations of liver**
 - Accessory liver
 - Alagille's syndrome
 - Congenital absence of liver
 - Congenital hepatomegaly
 - Congenital malformation of liver NOS

Q45 Other congenital malformations of digestive system

Excludes2: congenital diaphragmatic hernia (Q79.0)
congenital hiatus hernia (Q40.1)

- Q45.0 Agenesis, aplasia and hypoplasia of pancreas**
 - Congenital absence of pancreas
- Q45.1 Annular pancreas**
- Q45.2 Congenital pancreatic cyst**
- Q45.3 Other congenital malformations of pancreas and pancreatic duct**
 - Accessory pancreas
 - Congenital malformation of pancreas or pancreatic duct NOS
 - Excludes1: congenital diabetes mellitus (E10.-)
cystic fibrosis (E84.0-E84.9)
fibrocystic disease of pancreas (E84.-)
neonatal diabetes mellitus (P70.2)
- Q45.8 Other specified congenital malformations of digestive system**
 - Absence (complete) (partial) of alimentary tract NOS
 - Duplication of digestive system
 - Malposition, congenital of digestive system
- Q45.9 Congenital malformation of digestive system, unspecified**
 - Congenital anomaly of digestive system
 - Congenital deformity of digestive system

Congenital malformations of genital organs (Q50-Q56)

Excludes1: androgen resistance syndrome (E34.5)
syndromes associated with anomalies in the number and form of chromosomes
(Q90-Q99)
testicular feminization syndrome (E34.5)

Q50 Congenital malformations of ovaries, fallopian tubes and broad ligaments

Q50.0 Congenital absence of ovary

Excludes1: Turner's syndrome (Q96.-)

Q50.1 Developmental ovarian cyst

Q50.2 Congenital torsion of ovary

Q50.3 Other congenital malformations of ovary

Q50.31 Accessory ovary

Q50.32 Ovarian streak

46, XX with streak gonads

Q50.39 Other congenital malformation of ovary

Congenital malformation of ovary NOS

Q50.4 Embryonic cyst of fallopian tube

Fimbrial cyst

Q50.5 Embryonic cyst of broad ligament

Epoophoron cyst

Gartner's duct cyst

Parovarian cyst

Q50.6 Other congenital malformations of fallopian tube and broad ligament

Absence of fallopian tube and broad ligament

Accessory fallopian tube and broad ligament

Atresia of fallopian tube and broad ligament

Congenital malformation of fallopian tube or broad ligament NOS

Q51 Congenital malformations of uterus and cervix

Q51.0 Agenesis and aplasia of uterus

Congenital absence of uterus

Q51.1 Doubling of uterus with doubling of cervix and vagina

Q51.10 Doubling of uterus with doubling of cervix and vagina without obstruction

Doubling of uterus with doubling of cervix and vagina NOS

Q51.11 Doubling of uterus with doubling of cervix and vagina with obstruction

Q51.2 Other doubling of uterus

Doubling of uterus NOS

Q51.3 Bicornate uterus

Q51.4 Unicornate uterus

- Q51.5 Agenesis and aplasia of cervix**
Congenital absence of cervix
- Q51.6 Embryonic cyst of cervix**
- Q51.7 Congenital fistulae between uterus and digestive and urinary tracts**
- Q51.8 Other congenital malformations of uterus and cervix**
Hypoplasia of uterus and cervix
- Q51.9 Congenital malformation of uterus and cervix, unspecified**

Q52 Other congenital malformations of female genitalia

- Q52.0 Congenital absence of vagina**
- Q52.1 Doubling of vagina**
Septate vagina
Excludes1: doubling of vagina with doubling of uterus and cervix (Q51.1-)
- Q52.2 Congenital rectovaginal fistula**
Excludes1: cloaca (Q43.7)
- Q52.3 Imperforate hymen**
- Q52.4 Other congenital malformations of vagina**
Canal of Nuck cyst, congenital
Congenital malformation of vagina NOS
Embryonic vaginal cyst
- Q52.5 Fusion of labia**
- Q52.6 Congenital malformation of clitoris**
- Q52.7 Other and unspecified congenital malformations of vulva**
- Q52.70 Unspecified congenital malformations of vulva**
Congenital malformation of vulva NOS
- Q52.71 Congenital absence of vulva**
- Q52.79 Other congenital malformations of vulva**
Congenital cyst of vulva
- Q52.8 Other specified congenital malformations of female genitalia**
- Q52.9 Congenital malformation of female genitalia, unspecified**

Q53 Undescended and ectopic testicle

- Q53.0 Ectopic testis**
 - Q53.00 Ectopic testis, unspecified**
 - Q53.01 Ectopic testis, unilateral**
 - Q53.02 Ectopic testes, bilateral**
- Q53.1 Undescended testicle, unilateral**
 - Q53.10 Unspecified undescended testicle, unilateral**
 - Q53.11 Abdominal testis, unilateral**
 - Q53.12 Ectopic perineal testis, unilateral**
- Q53.2 Undescended testicle, bilateral**
 - Q53.20 Undescended testicle, unspecified, bilateral**
 - Q53.21 Abdominal testis, bilateral**
 - Q53.22 Ectopic perineal testis, bilateral**

Q53.9 Undescended testicle, unspecified
Cryptorchism NOS

Q54 Hypospadias

Excludes1: epispadias (Q64.0)

- Q54.0 Hypospadias, balanic**
Hypospadias, coronal
Hypospadias, glandular
- Q54.1 Hypospadias, penile**
- Q54.2 Hypospadias, penoscrotal**
- Q54.3 Hypospadias, perineal**
- Q54.4 Congenital chordee**
Chordee without hypospadias
- Q54.8 Other hypospadias**
Hypospadias with intersex state
- Q54.9 Hypospadias, unspecified**

Q55 Other congenital malformations of male genital organs

Excludes1: congenital hydrocele (P83.5)
hypospadias (Q54.-)

- Q55.0 Absence and aplasia of testis**
Monorchism
- Q55.1 Hypoplasia of testis and scrotum**
Fusion of testes
- Q55.2 Other and unspecified congenital malformations of testis and scrotum**
- Q55.20 Unspecified congenital malformations of testis and scrotum**
Congenital malformation of testis or scrotum NOS
- Q55.21 Polyorchism**
- Q55.22 Retractile testis**
- Q55.29 Other congenital malformations of testis and scrotum**
- Q55.3 Atresia of vas deferens**
Code first any associated cystic fibrosis (E84.-)
- Q55.4 Other congenital malformations of vas deferens, epididymis, seminal vesicles and prostate**
Absence or aplasia of prostate
Absence or aplasia of spermatic cord
Congenital malformation of vas deferens, epididymis, seminal vesicles or prostate NOS
- Q55.5 Congenital absence and aplasia of penis**
- Q55.6 Other congenital malformations of penis**
- Q55.61 Curvature of penis (lateral)**
- Q55.62 Hypoplasia of penis**
Micropenis
- Q55.69 Other congenital malformation of penis NOS**

- Congenital malformation of penis NOS
- Q55.7 Congenital vasocutaneous fistula**
- Q55.8 Other specified congenital malformations of male genital organs**
- Q55.9 Congenital malformation of male genital organ, unspecified**
- Congenital anomaly of male genital organ
- Congenital deformity of male genital organ

Q56 Indeterminate sex and pseudohermaphroditism

Excludes1: 46,XX true hermaphrodite (Q99.1)
chimera 46,XX/46,XY true hermaphrodite (Q99.0)
female pseudohermaphroditism with adrenocortical disorder (E25.-)
male pseudohermaphroditism with androgen resistance (E34.5)
pseudohermaphroditism with specified chromosomal anomaly (Q96-Q99)
pure gonadal dysgenesis (Q99.1)

- Q56.0 Hermaphroditism, not elsewhere classified**
Ovotestis
- Q56.1 Male pseudohermaphroditism, not elsewhere classified**
46, XY with streak gonads
Male pseudohermaphroditism NOS
- Q56.2 Female pseudohermaphroditism, not elsewhere classified**
Female pseudohermaphroditism NOS
- Q56.3 Pseudohermaphroditism, unspecified**
- Q56.4 Indeterminate sex, unspecified**
Ambiguous genitalia

Congenital malformations of the urinary system (Q60-Q64)

Q60 Renal agenesis and other reduction defects of kidney

Includes: congenital absence of kidney
congenital atrophy of kidney
infantile atrophy of kidney

- Q60.0 Renal agenesis, unilateral**
- Q60.1 Renal agenesis, bilateral**
- Q60.2 Renal agenesis, unspecified**
- Q60.3 Renal hypoplasia, unilateral**
- Q60.4 Renal hypoplasia, bilateral**
- Q60.5 Renal hypoplasia, unspecified**
- Q60.6 Potter's syndrome**

Q61 Cystic kidney disease

Excludes1: acquired cyst of kidney (N28.1)
Potter's syndrome (Q60.6)

- Q61.0 Congenital renal cyst
Q61.00 Congenital renal cyst, unspecified
 Cyst of kidney NOS (congenital)
Q61.01 Congenital single renal cyst
Q61.02 Congenital multiple renal cysts
- Q61.1 Polycystic kidney, infantile type
 Polycystic kidney, autosomal recessive
Q61.11 Cystic dilatation of collecting ducts
Q61.19 Other polycystic kidney, infantile type
- Q61.2 Polycystic kidney, adult type**
 Polycystic kidney, autosomal dominant
- Q61.3 Polycystic kidney, unspecified**
- Q61.4 Renal dysplasia**
- Q61.5 Medullary cystic kidney**
 Nephronophthisis
 Sponge kidney NOS
- Q61.8 Other cystic kidney diseases**
 Fibrocystic kidney
 Fibrocystic renal degeneration or disease
- Q61.9 Cystic kidney disease, unspecified**
 Meckel-Gruber syndrome

Q62 Congenital obstructive defects of renal pelvis and congenital malformations of ureter

- Q62.0 Congenital hydronephrosis**
- Q62.1 Congenital occlusion of ureter
 Atresia and stenosis of ureter
Q62.10 Congenital occlusion of ureter, unspecified
Q62.11 Congenital occlusion of ureteropelvic junction
Q62.12 Congenital occlusion of ureterovesical orifice
- Q62.2 Congenital megaureter**
 Congenital dilatation of ureter
- Q62.3 Other obstructive defects of renal pelvis and ureter
Q62.31 Congenital ureterocele, orthotopic
Q62.32 Cecoureterocele
 Ectopic ureterocele
Q62.39 Other obstructive defects of renal pelvis and ureter
- Q62.4 Agenesis of ureter**
 Congenital absence ureter
- Q62.5 Duplication of ureter**
 Accessory ureter
 Double ureter
- Q62.6 Malposition of ureter
Q62.60 Malposition of ureter, unspecified

- Q62.61 Deviation of ureter**
- Q62.62 Displacement of ureter**
- Q62.63 Anomalous implantation of ureter**
 - Ectopia of ureter
 - Ectopic ureter
- Q62.69 Other malposition of ureter**
- Q62.7 Congenital vesico-uretero-renal reflux**
- Q62.8 Other congenital malformations of ureter**
 - Anomaly of ureter NOS

Q63 Other congenital malformations of kidney

Excludes1: congenital nephrotic syndrome (N04.-)

- Q63.0 Accessory kidney**
- Q63.1 Lobulated, fused and horseshoe kidney**
- Q63.2 Ectopic kidney**
 - Congenital displaced kidney
 - Malrotation of kidney
- Q63.3 Hyperplastic and giant kidney**
 - Compensatory hypertrophy of kidney
- Q63.8 Other specified congenital malformations of kidney**
 - Congenital renal calculi
- Q63.9 Congenital malformation of kidney, unspecified**

Q64 Other congenital malformations of urinary system

- Q64.0 Epispadias**
 - Excludes1: hypospadias (Q54.-)
- Q64.1 Exstrophy of urinary bladder**
 - Q64.10 Exstrophy of urinary bladder, unspecified**
 - Ectopia vesicae
 - Q64.11 Supravesical fissure of urinary bladder**
 - Q64.12 Cloacal extrophy of urinary bladder**
 - Q64.19 Other exstrophy of urinary bladder**
 - Extroversion of bladder
- Q64.2 Congenital posterior urethral valves**
- Q64.3 Other atresia and stenosis of urethra and bladder neck**
 - Q64.31 Congenital bladder neck obstruction**
 - Congenital obstruction of vesicourethral orifice
 - Q64.32 Congenital stricture of urethra**
 - Q64.33 Congenital stricture of urinary meatus**
 - Q64.39 Other atresia and stenosis of urethra and bladder neck**
 - Atresia and stenosis of urethra and bladder neck NOS
- Q64.4 Malformation of urachus**
 - Cyst of urachus
 - Patent urachus

- Prolapse of urachus
- Q64.5 Congenital absence of bladder and urethra**
- Q64.6 Congenital diverticulum of bladder**
- Q64.7 Other and unspecified congenital malformations of bladder and urethra
Excludes1: congenital prolapse of bladder (mucosa) (Q79.4)
- Q64.70 Unspecified congenital malformation of bladder and urethra**
- Malformation of bladder or urethra NOS
- Q64.71 Congenital prolapse of urethra**
- Q64.72 Congenital prolapse of urinary meatus**
- Q64.73 Congenital urethrorectal fistula**
- Q64.74 Double urethra**
- Q64.75 Double urinary meatus**
- Q64.79 Other congenital malformations of bladder and urethra**
- Q64.8 Other specified congenital malformations of urinary system**
- Q64.9 Congenital malformation of urinary system, unspecified**
- Congenital anomaly NOS of urinary system
Congenital deformity NOS of urinary system

Congenital malformations and deformations of the musculoskeletal system (Q65-Q79)

Q65 Congenital deformities of hip

Excludes1: clicking hip (R29.4)

- Q65.0 Congenital dislocation of hip, unilateral
- Q65.00 Congenital dislocation of hip, unilateral, unspecified side**
- Q65.01 Congenital dislocation of right hip**
- Q65.02 Congenital dislocation of left hip**
- Q65.1 Congenital dislocation of hip, bilateral**
- Q65.2 Congenital dislocation of hip, unspecified**
- Q65.3 Congenital partial dislocation of hip, unilateral
- Q65.30 Congenital partial dislocation of hip, unilateral, unspecified side**
- Q65.31 Congenital partial dislocation of right hip**
- Q65.32 Congenital partial dislocation of left hip**
- Q65.4 Congenital partial dislocation of hip, bilateral**
- Q65.5 Congenital partial dislocation of hip, unspecified**
- Q65.6 Congenital unstable hip**
- Congenital dislocatable hip
- Q65.8 Other congenital deformities of hip**
- Anteversion of femoral neck
- Congenital acetabular dysplasia
- Congenital coxa valga
- Congenital coxa vara
- Q65.9 Congenital deformity of hip, unspecified**

Q66 Congenital deformities of feet

Excludes1: reduction defects of feet (Q72.-)
valgus deformities (acquired) (M21.0)
varus deformities (acquired) (M21.1)

Q66.0 Congenital talipes equinovarus

Q66.1 Congenital talipes calcaneovarus

Q66.2 Congenital metatarsus (primus) varus

Q66.3 Other congenital varus deformities of feet

Hallux varus, congenital

Q66.4 Congenital talipes calcaneovalgus

Q66.5 Congenital pes planus

Congenital flat foot

Congenital rigid flat foot

Congenital spastic (everted) flat foot

Excludes1: pes planus, acquired (M21.4)

Q66.6 Other congenital valgus deformities of feet

Congenital metatarsus valgus

Q66.7 Congenital pes cavus

Q66.8 Other congenital deformities of feet

Congenital asymmetric talipes

Congenital clubfoot NOS

Congenital talipes NOS

Congenital tarsal coalition

Congenital vertical talus

Hammer toe, congenital

Q66.9 Congenital deformity of feet, unspecified

Q67 Congenital musculoskeletal deformities of head, face, spine and chest

Excludes1: congenital malformation syndromes classified to Q87.-
Potter's syndrome (Q60.6)

Q67.0 Congenital facial asymmetry

Q67.1 Congenital compression facies

Q67.2 Dolichocephaly

Q67.3 Plagiocephaly

Q67.4 Other congenital deformities of skull, face and jaw

Congenital depressions in skull

Congenital hemifacial atrophy or hypertrophy

Deviation of nasal septum, congenital

Squashed or bent nose, congenital

Excludes1: dentofacial anomalies [including malocclusion] (M26-)

syphilitic saddle nose (A50.5)

Q67.5 Congenital deformity of spine
Congenital postural scoliosis
Congenital scoliosis NOS
Excludes1: infantile idiopathic scoliosis (M41.0)
scoliosis due to congenital bony malformation (Q76.3)

Q67.6 Pectus excavatum
Congenital funnel chest

Q67.7 Pectus carinatum
Congenital pigeon chest

Q67.8 Other congenital deformities of chest
Congenital deformity of chest wall NOS

Q68 Other congenital musculoskeletal deformities

Excludes1: reduction defects of limb(s) (Q71-Q73)

Q68.0 Congenital deformity of sternocleidomastoid muscle
Congenital contracture of sternocleidomastoid (muscle)
Congenital (sternomastoid) torticollis
Sternomastoid tumor (congenital)

Q68.1 Congenital deformity of hand
Congenital clubfinger
Spade-like hand (congenital)

Q68.2 Congenital deformity of knee
Congenital dislocation of knee
Congenital genu recurvatum

Q68.3 Congenital bowing of femur
Excludes1: anteverision of femur (neck) (Q65.8)

Q68.4 Congenital bowing of tibia and fibula

Q68.5 Congenital bowing of long bones of leg, unspecified

Q68.6 Discoid meniscus (congenital)
Q68.60 Discoid meniscus (congenital), unspecified meniscus
Q68.601 Discoid meniscus (congenital), unspecified meniscus, right knee
Q68.602 Discoid meniscus (congenital), unspecified meniscus, left knee
Q68.609 Discoid meniscus (congenital), unspecified meniscus, unspecified knee
Q68.61 Discoid meniscus (congenital), anterior horn of medial meniscus
Q68.611 Discoid meniscus (congenital), anterior horn of medial meniscus, right knee
Q68.612 Discoid meniscus (congenital), anterior horn of medial meniscus, left knee
Q68.619 Discoid meniscus (congenital), anterior horn of medial meniscus, unspecified knee
Q68.62 Discoid meniscus (congenital), posterior horn of medial meniscus
Q68.621 Discoid meniscus (congenital), posterior horn of medial

- meniscus, right knee
 - Q68.622 Discoid meniscus (congenital), posterior horn of medial meniscus, left knee**
 - Q68.629 Discoid meniscus (congenital), posterior horn of medial meniscus, unspecified knee**
 - Q68.63 Discoid meniscus (congenital), other medial meniscus
 - Discoid meniscus (congenital), medial meniscus NOS
 - Q68.631 Discoid meniscus (congenital), other medial meniscus, right knee**
 - Q68.632 Discoid meniscus (congenital), other medial meniscus, left knee**
 - Q68.639 Discoid meniscus (congenital), other medial meniscus, unspecified knee**
 - Q68.64 Discoid meniscus (congenital), anterior horn of lateral meniscus
 - Q68.641 Discoid meniscus (congenital), anterior horn of lateral meniscus, right knee**
 - Q68.642 Discoid meniscus (congenital), anterior horn of lateral meniscus, left knee**
 - Q68.649 Discoid meniscus (congenital), anterior horn of lateral meniscus, unspecified knee**
 - Q68.65 Discoid meniscus (congenital), posterior horn of lateral meniscus
 - Q68.651 Discoid meniscus (congenital), posterior horn of lateral meniscus, right knee**
 - Q68.652 Discoid meniscus (congenital), posterior horn of lateral meniscus, left knee**
 - Q68.659 Discoid meniscus (congenital), posterior horn of lateral meniscus, unspecified knee**
 - Q68.66 Discoid meniscus (congenital), other lateral meniscus
 - Discoid meniscus (congenital), lateral meniscus NOS
 - Q68.661 Discoid meniscus (congenital), other lateral meniscus, right knee**
 - Q68.662 Discoid meniscus (congenital), other lateral meniscus, left knee**
 - Q68.669 Discoid meniscus (congenital), other lateral meniscus, unspecified knee**
- Q68.8 Other specified congenital musculoskeletal deformities**
- Congenital deformity of clavicle
 - Congenital deformity of elbow
 - Congenital deformity of forearm
 - Congenital deformity of scapula
 - Congenital dislocation of elbow
 - Congenital dislocation of shoulder

Q69 Polydactyly

Q69.0 Accessory finger(s)

- Q69.1 Accessory thumb(s)**
- Q69.2 Accessory toe(s)**
 - Accessory hallux
- Q69.9 Polydactyly, unspecified**
 - Supernumerary digit(s) NOS

Q70 Syndactyly

- Q70.0 Fused fingers**
 - Complex syndactyly of fingers with synostosis
- Q70.00 Fused fingers, unspecified fingers**
- Q70.01 Fused right fingers**
- Q70.02 Fused left fingers**
- Q70.03 Fused fingers, bilateral**
- Q70.1 Webbed fingers**
 - Simple syndactyly of fingers without synostosis
- Q70.10 Webbed fingers, unspecified side**
- Q70.11 Webbed right fingers**
- Q70.12 Webbed left fingers**
- Q70.13 Webbed fingers, bilateral**
- Q70.2 Fused toes**
 - Complex syndactyly of toes with synostosis
- Q70.3 Webbed toes**
 - Simple syndactyly of toes without synostosis
- Q70.4 Polysyndactyly**
- Q70.9 Syndactyly, unspecified**
 - Symphalangy NOS

Q71 Reduction defects of upper limb

- Q71.0 Congenital complete absence of upper limb**
 - Q71.00 Congenital complete absence of upper limb, unspecified side**
 - Q71.01 Congenital complete absence of right upper limb**
 - Q71.02 Congenital complete absence of left upper limb**
 - Q71.03 Congenital complete absence of upper limb, bilateral**
- Q71.1 Congenital absence of upper arm and forearm with hand present**
 - Q71.10 Congenital absence of upper arm and forearm with hand present, unspecified side**
 - Q71.11 Congenital absence of right upper arm and forearm with hand present**
 - Q71.12 Congenital absence of left upper arm and forearm with hand present**
 - Q71.13 Congenital absence of upper arm and forearm with hand present, bilateral**
- Q71.2 Congenital absence of both forearm and hand**
 - Q71.20 Congenital absence of both forearm and hand, unspecified side**

- Q71.21** Congenital absence of both right forearm and hand
 - Q71.22** Congenital absence of both left forearm and hand
 - Q71.23** Congenital absence of both forearm and hand, bilateral
- Q71.3 Congenital absence of hand and finger
 - Q71.30** Congenital absence of hand and finger, unspecified side
 - Q71.31** Congenital absence of right hand and finger
 - Q71.32** Congenital absence of left hand and finger
 - Q71.33** Congenital absence of hand and finger, bilateral
- Q71.4 Longitudinal reduction defect of radius
 - Clubhand (congenital)
 - Radial clubhand
 - Q71.40** Longitudinal reduction defect of radius, unspecified side
 - Q71.41** Longitudinal reduction defect of right radius
 - Q71.42** Longitudinal reduction defect of left radius
 - Q71.43** Longitudinal reduction defect of radius, bilateral
- Q71.5 Longitudinal reduction defect of ulna
 - Q71.50** Longitudinal reduction defect of ulna, unspecified side
 - Q71.51** Longitudinal reduction defect of right ulna
 - Q71.52** Longitudinal reduction defect of left ulna
 - Q71.53** Longitudinal reduction defect of ulna, bilateral
- Q71.6 Lobster-claw hand
 - Q71.60** Lobster-claw hand, unspecified side
 - Q71.61** Lobster-claw right hand
 - Q71.62** Lobster-claw left hand
 - Q71.63** Lobster-claw hand, bilateral
- Q71.8 Other reduction defects of upper limb
 - Congenital shortening of upper limb
 - Q71.80** Other reduction defects of upper limb, unspecified side
 - Q71.81** Other reduction defects of right upper limb
 - Q71.82** Other reduction defects of left upper limb
 - Q71.83** Other reduction defects of upper limb, bilateral
- Q71.9 Reduction defect of upper limb, unspecified
 - Q71.90** Reduction defect of upper limb, unspecified, side unspecified
 - Q71.91** Reduction defect of right upper limb, unspecified
 - Q71.92** Reduction defect of left upper limb, unspecified
 - Q71.93** Reduction defect of upper limb, unspecified, bilateral

Q72 Reduction defects of lower limb

- Q72.0 Congenital complete absence of lower limb
 - Q72.00** Congenital complete absence of lower limb, unspecified side
 - Q72.01** Congenital complete absence of right lower limb
 - Q72.02** Congenital complete absence of left lower limb
 - Q72.03** Congenital complete absence of lower limb, bilateral
- Q72.1 Congenital absence of thigh and lower leg with foot present
 - Q72.10** Congenital absence of thigh and lower leg with foot present,

- unspecified side
- Q72.11 Congenital absence of right thigh and lower leg with foot present
- Q72.12 Congenital absence of left thigh and lower leg with foot present
- Q72.13 Congenital absence of thigh and lower leg with foot present, bilateral
- Q72.2 Congenital absence of both lower leg and foot
- Q72.20 Congenital absence of both lower leg and foot, unspecified side
- Q72.21 Congenital absence of both right lower leg and foot
- Q72.22 Congenital absence of both left lower leg and foot
- Q72.23 Congenital absence of both lower leg and foot, bilateral
- Q72.3 Congenital absence of foot and toe(s)
- Q72.30 Congenital absence of foot and toe(s), unspecified side
- Q72.31 Congenital absence of right foot and toe(s)
- Q72.32 Congenital absence of left foot and toe(s)
- Q72.33 Congenital absence of foot and toe(s), bilateral
- Q72.4 Longitudinal reduction defect of femur
- Proximal femoral focal deficiency
- Q72.40 Longitudinal reduction defect of femur, unspecified side
- Q72.41 Longitudinal reduction defect of right femur
- Q72.42 Longitudinal reduction defect of left femur
- Q72.43 Longitudinal reduction defect of femur, bilateral
- Q72.5 Longitudinal reduction defect of tibia
- Q72.50 Longitudinal reduction defect of tibia, unspecified side
- Q72.51 Longitudinal reduction defect of right tibia
- Q72.52 Longitudinal reduction defect of left tibia
- Q72.53 Longitudinal reduction defect of tibia, bilateral
- Q72.6 Longitudinal reduction defect of fibula
- Q72.60 Longitudinal reduction defect of fibula, unspecified side
- Q72.61 Longitudinal reduction defect of right fibula
- Q72.62 Longitudinal reduction defect of left fibula
- Q72.63 Longitudinal reduction defect of fibula, bilateral
- Q72.7 Split foot
- Q72.70 Split foot, unspecified side
- Q72.71 Right split foot
- Q72.72 Left split foot
- Q72.73 Split foot, bilateral
- Q72.8 Other reduction defects of lower limb
- Congenital shortening of lower limb(s)
- Q72.80 Other reduction defects of lower limb, unspecified side
- Q72.81 Other reduction defects of right lower limb
- Q72.82 Other reduction defects of left lower limb
- Q72.83 Other reduction defects of lower limb, bilateral
- Q72.9 Reduction defect of lower limb, unspecified
- Q72.90 Reduction defect of lower limb, unspecified, unspecified side
- Q72.91 Reduction defect of right lower limb, unspecified
- Q72.92 Reduction defect of left lower limb, unspecified

Q72.93 Reduction defect of lower limb, unspecified, bilateral

Q73 Reduction defects of unspecified limb

- Q73.0 Congenital absence of unspecified limb(s)**
Amelia NOS
- Q73.1 Phocomelia, unspecified limb(s)**
Phocomelia NOS
- Q73.8 Other reduction defects of unspecified limb(s)**
Longitudinal reduction deformity of unspecified limb(s)
Ectromelia of limb NOS
Hemimelia of limb NOS
Reduction defect of limb NOS

Q74 Other congenital malformations of limb(s)

Excludes1: polydactyly (Q69.-)
reduction defect of limb (Q71-Q73)
syndactyly (Q70.-)

- Q74.0 Other congenital malformations of upper limb(s), including shoulder girdle**
Accessory carpal bones
Cleidocranial dysostosis
Congenital pseudarthrosis of clavicle
Macrodactyly (fingers)
Madelung's deformity
Radioulnar synostosis
Sprengel's deformity
Triphalangeal thumb
- Q74.1 Congenital malformation of knee**
Congenital absence of patella
Congenital dislocation of patella
Congenital genu valgum
Congenital genu varum
Rudimentary patella
Excludes1: congenital dislocation of knee (Q68.2)
congenital genu recurvatum (Q68.2)
nail patella syndrome (Q87.2)
- Q74.2 Other congenital malformations of lower limb(s), including pelvic girdle**
Congenital fusion of sacroiliac joint
Congenital malformation of ankle joint
Congenital malformation of sacroiliac joint
Excludes1: anteversion of femur (neck) (Q65.8)
- Q74.3 Arthrogryposis multiplex congenita**
- Q74.8 Other specified congenital malformations of limb(s)**
- Q74.9 Unspecified congenital malformation of limb(s)**
Congenital anomaly of limb(s) NOS

Q75 Other congenital malformations of skull and face bones

Excludes1: congenital malformation of face NOS (Q18.-)
congenital malformation syndromes classified to Q87.-
dentofacial anomalies [including malocclusion] (M26.-)
musculoskeletal deformities of head and face (Q67.0-Q67.4)
skull defects associated with congenital anomalies of brain such as:
anencephaly (Q00.0)
encephalocele (Q01.-)
hydrocephalus (Q03.-)
microcephaly (Q02)

Q75.0 Craniosynostosis

Acrocephaly
Imperfect fusion of skull
Oxycephaly
Trigonocephaly

Q75.1 Craniofacial dysostosis

Crouzon's disease

Q75.2 Hypertelorism

Q75.3 Macrocephaly

Q75.4 Mandibulofacial dysostosis

Q75.5 Oculomandibular dysostosis

Q75.8 Other specified congenital malformations of skull and face bones

Absence of skull bone, congenital
Congenital deformity of forehead
Platybasia

Q75.9 Congenital malformation of skull and face bones, unspecified

Congenital anomaly of face bones NOS
Congenital anomaly of skull NOS

Q76 Congenital malformations of spine and bony thorax

Excludes1: congenital musculoskeletal deformities of spine and chest (Q67.5-Q67.8)

Q76.0 Spina bifida occulta

Excludes1: meningocele (spinal) (Q05.-)
spina bifida (aperta) (cystica) (Q05.-)

Q76.1 Klippel-Feil syndrome

Cervical fusion syndrome

Q76.2 Congenital spondylolisthesis

Congenital spondylolisthesis

Excludes1: spondylolisthesis (acquired) (M43.1-)
spondylolisthesis (acquired) (M43.0-)

Q76.3 Congenital scoliosis due to congenital bony malformation

Hemivertebra fusion or failure of segmentation with scoliosis

Q76.4 Other congenital malformations of spine, not associated with scoliosis

Q76.41 Congenital kyphosis

- Q76.411** Congenital kyphosis, occipito-atlanto-axial region
- Q76.412** Congenital kyphosis, cervical region
- Q76.413** Congenital kyphosis, cervicothoracic region
- Q76.414** Congenital kyphosis, thoracic region
- Q76.415** Congenital kyphosis, thoracolumbar region
- Q76.419** Congenital kyphosis, unspecified region
- Q76.42** Congenital lordosis
 - Q76.425** Congenital lordosis, thoracolumbar region
 - Q76.426** Congenital lordosis, lumbar region
 - Q76.427** Congenital lordosis, lumbosacral region
 - Q76.428** Congenital lordosis, sacral and sacrococcygeal region
 - Q76.429** Congenital lordosis, unspecified region
- Q76.49** Other congenital malformations of spine, not associated with scoliosis
 - Congenital absence of vertebra NOS
 - Congenital fusion of spine NOS
 - Congenital malformation of lumbosacral (joint) (region) NOS
 - Congenital malformation of spine NOS
 - Hemivertebra NOS
 - Malformation of spine NOS
 - Platyspondylisis NOS
 - Supernumerary vertebra NOS
- Q76.5** Cervical rib
 - Supernumerary rib in cervical region
- Q76.6** Other congenital malformations of ribs
 - Accessory rib
 - Congenital absence of rib
 - Congenital fusion of ribs
 - Congenital malformation of ribs NOS
 - Excludes1: short rib syndrome (Q77.2)
- Q76.7** Congenital malformation of sternum
 - Congenital absence of sternum
 - Sternum bifidum
- Q76.8** Other congenital malformations of bony thorax
- Q76.9** Congenital malformation of bony thorax, unspecified

Q77 Osteochondrodysplasia with defects of growth of tubular bones and spine

Excludes1: mucopolysaccharidosis (E76.0-E76.3)

- Q77.0** Achondrogenesis
 - Hypochondrogenesis
- Q77.1** Thanatophoric short stature
- Q77.2** Short rib syndrome
 - Asphyxiating thoracic dysplasia [Jeune]
- Q77.3** Chondrodysplasia punctata

- Excludes1: Rhizomelic chondrodysplasia punctata (E71.430)
- Q77.4 Achondroplasia**
Hypochondroplasia
- Q77.5 Diastrophic dysplasia**
- Q77.6 Chondroectodermal dysplasia**
Ellis-van Creveld syndrome
- Q77.7 Spondyloepiphyseal dysplasia**
- Q77.8 Other osteochondrodysplasia with defects of growth of tubular bones and spine**
- Q77.9 Osteochondrodysplasia with defects of growth of tubular bones and spine, unspecified**

Q78 Other osteochondrodysplasias

- Q78.0 Osteogenesis imperfecta**
Fragilitas ossium
Osteopsathyrosis
- Q78.1 Polyostotic fibrous dysplasia**
Albright(-McCune)(-Sternberg) syndrome
- Q78.2 Osteopetrosis**
Albers-Schönberg syndrome
- Q78.3 Progressive diaphyseal dysplasia**
Camurati-Engelmann syndrome
- Q78.4 Enchondromatosis**
Maffucci's syndrome
Ollier's disease
- Q78.5 Metaphyseal dysplasia**
Pyle's syndrome
- Q78.6 Multiple congenital exostoses**
Diaphyseal aclasis
- Q78.8 Other specified osteochondrodysplasias**
Osteopoikilosis
- Q78.9 Osteochondrodysplasia, unspecified**
Chondrodystrophy NOS
Osteodystrophy NOS

Q79 Congenital malformations of musculoskeletal system, not elsewhere classified

Excludes2: congenital (sternomastoid) torticollis (Q68.0)

- Q79.0 Congenital diaphragmatic hernia**
Excludes1: congenital hiatus hernia (Q40.1)
- Q79.1 Other congenital malformations of diaphragm**
Absence of diaphragm
Congenital malformation of diaphragm NOS

- Eventration of diaphragm
- Q79.2 Exomphalos**
Omphalocele
Excludes1: umbilical hernia (K42.-)
- Q79.3 Gastroschisis**
- Q79.4 Prune belly syndrome**
Congenital prolapse of bladder mucosa
Eagle-Barrett syndrome
- Q79.5 Other congenital malformations of abdominal wall**
Excludes1: umbilical hernia (K42.-)
- Q79.51 Congenital hernia of bladder**
- Q79.59 Other congenital malformations of abdominal wall**
- Q79.6 Ehlers-Danlos syndrome**
- Q79.8 Other congenital malformations of musculoskeletal system**
Absence of muscle
Absence of tendon
Accessory muscle
Amyotrophy congenita
Congenital constricting bands
Congenital shortening of tendon
Poland's syndrome
- Q79.9 Congenital malformation of musculoskeletal system, unspecified**
Congenital anomaly of musculoskeletal system NOS
Congenital deformity of musculoskeletal system NOS

Other congenital malformations (Q80-Q89)

Q80 Congenital ichthyosis

Excludes1: Refsum's disease (G60.1)

- Q80.0 Ichthyosis vulgaris**
- Q80.1 X-linked ichthyosis**
- Q80.2 Lamellar ichthyosis**
Collodion baby
- Q80.3 Congenital bullous ichthyosiform erythroderma**
- Q80.4 Harlequin fetus**
- Q80.8 Other congenital ichthyosis**
- Q80.9 Congenital ichthyosis, unspecified**

Q81 Epidermolysis bullosa

Q81.0 Epidermolysis bullosa simplex

Excludes1: Cockayne's syndrome (Q87.1)

- Q81.1 Epidermolysis bullosa letalis**
Herlitz' syndrome
- Q81.2 Epidermolysis bullosa dystrophica**
- Q81.8 Other epidermolysis bullosa**
- Q81.9 Epidermolysis bullosa, unspecified**

Q82 Other congenital malformations of skin

Excludes1: acrodermatitis enteropathica (E83.2)
congenital erythropoietic porphyria (E80.0)
pilonidal cyst or sinus (L05.-)
Sturge-Weber (-Dimitri) syndrome (Q85.8)

- Q82.0 Hereditary lymphedema**
- Q82.1 Xeroderma pigmentosum**
- Q82.2 Mastocytosis**
Urticaria pigmentosa
Excludes1: malignant mastocytosis (C96.2)
- Q82.3 Incontinentia pigmenti**
- Q82.4 Ectodermal dysplasia (anhidrotic)**
Excludes1: Ellis-van Creveld syndrome (Q77.6)
- Q82.5 Congenital non-neoplastic nevus**

Birthmark NOS
Flammeus Nevus
Portwine Nevus
Sanguineous Nevus
Strawberry Nevus
Vascular Nevus NOS
Verrucous Nevus
Excludes1: café au lait spots (L81.3)
lentigo (L81.4)
nevus NOS (D22.-)
araneus nevus (I78.1)
melanocytic nevus (D22.-)
pigmented nevus (D22.-)
spider nevus (I78.1)
stellar nevus (I78.1)

Q82.8 Other specified congenital malformations of skin

Abnormal palmar creases
Accessory skin tags
Benign familial pemphigus [Hailey-Hailey]
Congenital poikiloderma
Cutis laxa (hyperelastica)
Dermatoglyphic anomalies
Inherited keratosis palmaris et plantaris
Keratosis follicularis [Darier-White]
Excludes1: Ehlers-Danlos syndrome (Q79.6)

Q82.9 Congenital malformation of skin, unspecified

Q83 Congenital malformations of breast

Excludes2: absence of pectoral muscle (Q79.8)

Q83.0 Congenital absence of breast with absent nipple

Q83.1 Accessory breast

Supernumerary breast

Q83.2 Absent nipple

Q83.3 Accessory nipple

Supernumerary nipple

Q83.8 Other congenital malformations of breast

Hypoplasia of breast

Q83.9 Congenital malformation of breast, unspecified

Q84 Other congenital malformations of integument

Q84.0 Congenital alopecia

Congenital atrichosis

Q84.1 Congenital morphological disturbances of hair, not elsewhere classified

Beaded hair

Monilethrix

Pili annulati

Excludes1: Menkes' kinky hair syndrome (E83.0)

Q84.2 Other congenital malformations of hair

Congenital hypertrichosis

Congenital malformation of hair NOS

Persistent lanugo

Q84.3 Anonychia

Excludes1: nail patella syndrome (Q87.2)

Q84.4 Congenital leukonychia

Q84.5 Enlarged and hypertrophic nails

Congenital onychauxis

Pachyonychia

Q84.6 Other congenital malformations of nails

Congenital clubnail

Congenital koilonychia

Congenital malformation of nail NOS

Q84.8 Other specified congenital malformations of integument

Aplasia cutis congenita

Q84.9 Congenital malformation of integument, unspecified

Congenital anomaly of integument NOS

Congenital deformity of integument NOS

Q85 Phakomatoses, not elsewhere classified

Excludes1: ataxia telangiectasia [Louis-Bar] (G11.3)
familial dysautonomia [Riley-Day] (G90.1)

Q85.0 Neurofibromatosis (nonmalignant)

Von Recklinghausen's disease

Q85.1 Tuberous sclerosis

Bourneville's disease

Epiloia

Q85.8 Other phakomatoses, not elsewhere classified

Peutz-Jeghers Syndrome

Sturge-Weber(-Dimitri) syndrome

von Hippel-Lindau syndrome

Excludes1: Meckel-Gruber syndrome (Q61.9)

Q85.9 Phakomatosis, unspecified

Hamartosis NOS

Q86 Congenital malformation syndromes due to known exogenous causes, not elsewhere classified

Excludes2: iodine-deficiency-related hypothyroidism (E00-E02)

nonteratogenic effects of substances transmitted via placenta or breast milk
(P04.-)

Q86.0 Fetal alcohol syndrome (dysmorphic)

Q86.1 Fetal hydantoin syndrome

Meadow's syndrome

Q86.2 Dysmorphism due to Warfarin

Q86.8 Other congenital malformation syndromes due to known exogenous causes

Q87 Other specified congenital malformation syndromes affecting multiple systems

Use additional code(s) to identify all associated manifestations

Q87.0 Congenital malformation syndromes predominantly affecting facial appearance

Acrocephalopolysyndactyly

Acrocephalosyndactyly [Apert]

Cryptophthalmos syndrome

Cyclopia

Goldenhar syndrome

Moebius syndrome

Oro-facial-digital syndrome

Robin syndrome

Treacher Collins syndrome

Whistling face

Q87.1 Congenital malformation syndromes predominantly associated with short

stature

Aarskog syndrome
Cockayne syndrome
De Lange syndrome
Dubowitz syndrome
Noonan syndrome
Prader-Willi syndrome
Robinow-Silverman-Smith syndrome
Russell-Silver syndrome
Seckel syndrome
Smith-Lemli-Opitz syndrome

Excludes1: Ellis-van Creveld syndrome (Q77.6)

Q87.2 Congenital malformation syndromes predominantly involving limbs

Holt-Oram syndrome
Klippel-Trenaunay-Weber syndrome
Nail patella syndrome
Rubinstein-Taybi syndrome
Sirenomelia syndrome
Thrombocytopenia with absent radius [TAR] syndrome
VATER syndrome

Q87.3 Congenital malformation syndromes involving early overgrowth

Beckwith-Wiedemann syndrome
Sotos' syndrome
Weaver syndrome

Q87.4 Marfan's syndrome

Q87.40 Marfan's syndrome, unspecified

Q87.41 Marfan's syndrome with cardiovascular manifestations

Q87.410 Marfan's syndrome with aortic dilation

Q87.418 Marfan's syndrome with other cardiovascular manifestations

Q87.42 Marfan's syndrome with ocular manifestations

Q87.43 Marfan's syndrome with skeletal manifestation

Q87.5 Other congenital malformation syndromes with other skeletal changes

Q87.8 Other specified congenital malformation syndromes, not elsewhere classified

Q87.81 Alport syndrome

Q87.810 Alport syndrome without chronic renal failure

Alport syndrome NOS

Q87.811 Alport syndrome with chronic renal failure

Q87.89 Other specified congenital malformation syndromes, not elsewhere classified

Laurence-Moon (-Bardet)-Biedl syndrome

Q89 Other congenital malformations, not elsewhere classified

Q89.0 Congenital absence and malformations of spleen

Excludes1: isomerism of atrial appendages (with asplenia or polysplenia)

(Q20.6)

Q89.01 Asplenia (congenital)

Q89.09 Congenital malformations of spleen

Congenital splenomegaly

Q89.1 Congenital malformations of adrenal gland

Excludes1: adrenogenital disorders (E25.-)

congenital adrenal hyperplasia (E25.0)

Q89.2 Congenital malformations of other endocrine glands

Congenital malformation of parathyroid or thyroid gland

Persistent thyroglossal duct

Thyroglossal cyst

Excludes1: congenital goiter (E03.0)

congenital hypothyroidism (E03.1)

Q89.3 Situs inversus

Dextrocardia with situs inversus

Mirror-image atrial arrangement with situs inversus

Situs inversus or transversus abdominalis

Situs inversus or transversus thoracis

Transposition of abdominal viscera

Transposition of thoracic viscera

Excludes1: dextrocardia NOS (Q24.0)

Q89.4 Conjoined twins

Craniopagus

Dicephaly

Pygopagus

Thoracopagus

Q89.7 Multiple congenital malformations, not elsewhere classified

Multiple congenital anomalies NOS

Multiple congenital deformities NOS

Excludes1: congenital malformation syndromes affecting multiple systems
(Q87.-)

Q89.8 Other specified congenital malformations

Q89.9 Congenital malformation, unspecified

Congenital anomaly NOS

Congenital deformity NOS

Chromosomal abnormalities, not elsewhere classified

(Q90-Q99)

Q90 Down syndrome

Use additional code(s) to identify any associated physical conditions

Q90.0 Trisomy 21, nonmosaicism (meiotic nondisjunction)

Q90.1 Trisomy 21, mosaicism (mitotic nondisjunction)

Q90.2 Trisomy 21, translocation

Q90.9 Down's syndrome, unspecified
Trisomy 21 NOS

Q91 Trisomy 18 and Trisomy 13

- Q91.0 Trisomy 18, nonmosaicism (meiotic nondisjunction)**
- Q91.1 Trisomy 18, mosaicism (mitotic nondisjunction)**
- Q91.2 Trisomy 18, translocation**
- Q91.3 Trisomy 18, unspecified**
- Q91.4 Trisomy 13, nonmosaicism (meiotic nondisjunction)**
- Q91.5 Trisomy 13, mosaicism (mitotic nondisjunction)**
- Q91.6 Trisomy 13, translocation**
- Q91.7 Trisomy 13, unspecified**

Q92 Other trisomies and partial trisomies of the autosomes, not elsewhere classified

Includes: unbalanced translocations and insertions

Excludes1: trisomies of chromosomes 13, 18, 21 (Q90-Q91)

- Q92.0 Whole chromosome trisomy, nonmosaicism (meiotic nondisjunction)**
- Q92.1 Whole chromosome trisomy, mosaicism (mitotic nondisjunction)**
- Q92.2 Partial trisomy**
 - Less than whole arm duplicated
 - Whole arm or more duplicated
- Excludes1: partial trisomy due to unbalanced translocation (Q92.5)
- Q92.5 Duplications with other complex rearrangements**
 - Partial trisomy due to unbalanced translocations
 - Code also any associated deletions due to unbalanced translocations, inversions and insertions (Q93.7)
- Q92.6 Marker chromosomes**
 - Trisomies due to dicentrics
 - Trisomies due to extra rings
 - Trisomies due to isochromosomes
 - Individual with marker heterochromatin
- Q92.61 Marker chromosomes in normal individual**
- Q92.62 Marker chromosomes in abnormal individual**
- Q92.7 Triploidy and polyploidy**
- Q92.8 Other specified trisomies and partial trisomies of autosomes**
 - Duplications identified by fluorescence in situ hybridization (FISH)
 - Duplications identified by in situ hybridization (ISH)
 - Duplications seen only at prometaphase
- Q92.9 Trisomy and partial trisomy of autosomes, unspecified**

Q93 Monosomies and deletions from the autosomes, not elsewhere classified

- Q93.0** Whole chromosome monosomy, nonmosaicism (meiotic nondisjunction)
Q93.1 Whole chromosome monosomy, mosaicism (mitotic nondisjunction)
Q93.2 Chromosome replaced with ring, dicentric or isochromosome
Q93.3 Deletion of short arm of chromosome 4
Wolff-Hirschorn syndrome
Q93.4 Deletion of short arm of chromosome 5
Cri-du-chat syndrome
Q93.7 Deletions with other complex rearrangements
Deletions due to unbalanced translocations, inversions and insertions
Code also any associated duplications due to unbalanced translocations, inversions and insertions (Q92.5)
Q93.8 Other deletions from the autosomes
Deletions identified by fluorescence in situ hybridization (FISH)
Deletions identified by in situ hybridization (ISH)
Deletions seen only at prometaphase
Q93.9 Deletion from autosomes, unspecified

Q95 Balanced rearrangements and structural markers, not elsewhere classified

Includes: Robertsonian and balanced reciprocal translocations and insertions

- Q95.0** Balanced translocation and insertion in normal individual
Q95.1 Chromosome inversion in normal individual
Q95.2 Balanced autosomal rearrangement in abnormal individual
Q95.3 Balanced sex/autosomal rearrangement in abnormal individual
Q95.5 Individual with autosomal fragile site
Q95.8 Other balanced rearrangements and structural markers
Q95.9 Balanced rearrangement and structural marker, unspecified

Q96 Turner syndrome

Excludes1: Noonan syndrome (Q87.1)

- Q96.0** Karyotype 45, X
Q96.1 Karyotype 46, X i(Xq)
Karyotype 46, isochromosome Xq
Q96.2 Karyotype 46, X with abnormal sex chromosome, except i(Xq)
Karyotype 46, X with abnormal sex chromosome, except isochromosome Xq
Q96.3 Mosaicism, 45, X/46, XX or XY
Q96.4 Mosaicism, 45, X/other cell line(s) with abnormal sex chromosome
Q96.8 Other variants of Turner syndrome
Q96.9 Turner syndrome, unspecified

Q97 Other sex chromosome abnormalities, female phenotype, not elsewhere classified

Excludes1: Turner syndrome (Q96.-)

- Q97.0 Karyotype 47, XXX**
- Q97.1 Female with more than three X chromosomes**
- Q97.2 Mosaicism, lines with various numbers of X chromosomes**
- Q97.3 Female with 46, XY karyotype**
- Q97.8 Other specified sex chromosome abnormalities, female phenotype**
- Q97.9 Sex chromosome abnormality, female phenotype, unspecified**

Q98 Other sex chromosome abnormalities, male phenotype, not elsewhere classified

- Q98.0 Klinefelter syndrome karyotype 47, XXY**
- Q98.1 Klinefelter syndrome, male with more than two X chromosomes**
- Q98.3 Male with 46, XX karyotype**
- Q98.4 Klinefelter syndrome, unspecified**
- Q98.5 Karyotype 47, XYY**
- Q98.6 Male with structurally abnormal sex chromosome**
- Q98.7 Male with sex chromosome mosaicism**
- Q98.8 Other specified sex chromosome abnormalities, male phenotype**
- Q98.9 Sex chromosome abnormality, male phenotype, unspecified**

Q99 Other chromosome abnormalities, not elsewhere classified

- Q99.0 Chimera 46, XX/46, XY**
Chimera 46, XX/46, XY true hermaphrodite
- Q99.1 46, XX true hermaphrodite**
 - 46, XX with streak gonads
 - 46, XY with streak gonads
 - Pure gonadal dysgenesis
- Q99.2 Fragile X chromosome**
Fragile X syndrome
- Q99.8 Other specified chromosome abnormalities**
- Q99.9 Chromosomal abnormality, unspecified**